CIDP misdiagnosis: a clinical more than electrophysiogical problem?

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LETTER

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"We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines."

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I read with great interest the paper by Allen et al. on the contribution of electrodiagnostic errors to the misdiagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP) (1).

The authors retrospectively reviewed data from 86 patients who had received the diagnosis and concluded that 39 (45%) did not have CIDP. Amongst those, data were analyzed from 29.

Thirteen (45%) were found to have initially correctly-interpreted electrophysiological data, but only one, with CMT4C, demonstrated demyelinating electrophysiology. The authors do not detail whether this patient had, albeit asymmetrical, proximal weakness which may explain a misdiagnosis or concurrent diagnosis of CIDP (2). The other patients surprisingly had normal results, axonopathy or motor neuron disease.

Misinterpreted initial studies with reported “demyelinating” or “mixed axonal and demyelinating” findings were observed in 16 subjects. Although similarly due to various reasons and different final diagnoses (axonal neuropathy, compressive neuropathy, motor neuron disease or even normal), none, in short, met minimal EFNS/PNS electrodiagnostic requirements for demyelination (3).

Electrodiagnostic misinterpretation is an important issue to consider in CIDP misdiagnosis but as illustrated in this study, may occur when subjective interpretations are made without regard for existing highly sensitive and specific criteria that are clearly described and easy to apply (4). This would therefore appear readily remediable if guidelines were simply used.

The authors’ recommendation regarding neurophysiology training programmes is appropriate but the fact they found that nearly half of misdiagnosed cases actually had a correctly-interpreted study, raises the issue of clinical diagnosis as a fundamental problem. With regard to the incorrectly-interpreted electrodiagnostic studies, the presence of motor neuron disease,

and even more so, of small fibre neuropathy or no neuropathy, equally enhances this concern regarding the clinical diagnosis/suspicion, which is hard to reconcile with CIDP. In these patients, the question is how *any* electrophysiological interpretation could possibly have suggested CIDP?

Of note, a minority (30.5%) of misdiagnosed cases met clinical EFNS/PNS criteria for "atypical CIDP", and none for "typical CIDP". "Atypical CIDP" represents a very diverse group (3), which may occasionally lead to erroneous diagnostic considerations. However, although the temptation to attempt treatment in selected cases may be at times understandable, clinical judgement is essential to differentiate for example, motor neuron disease from possible Lewis-Sumner syndrome, or length-dependent axonal neuropathy from possible sensory ataxic CIDP. That 69.5% of misdiagnosed cases did not meet any clinical criteria for CIDP is perplexing and highly concerning.

The authors rightly make several important suggestions to eradicate electrophysiological misdiagnosis but, for a start, clinical findings need to take priority over any test results, including electrophysiology or mildly abnormal cerebrospinal fluid protein levels. Reliance on subjective amelioration post-treatment, as demonstrated in the authors' previous work (5), also represents a serious clinical failure, likely as, if not more, problematic than electrophysiology, because it may result in unnecessarily prolonged treatment.

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